Functional outcome after complete surgical removal of giant vestibular schwannomas

Clinical article

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Object. The authors evaluated the outcome of radical surgery in a consecutive series of patients with giant vestibular schwannomas (VSs).

Methods. Fifty patients with VSs > 4.0 cm in maximal extrameatal diameter were included in this retrospective study (Group A). The group was compared with a matched group of 167 patients with VSs < 3.9 cm (Group B). In all cases the retrosigmoid approach was used. Outcome measures included completeness of tumor removal, facial nerve function, hearing, and the surgery-related complication rate.

Results. The mean tumor size in Group A was 4.4 cm and that in Group B was 2.3 cm. Total removal was achieved in all Group A patients and in 97.6% of Group B patients. The anatomical integrity of the facial nerve was preserved in 92% in Group A and in 98.8% in Group B. At last follow-up 75% of the patients with giant VSs had excellent or good facial nerve function, 19% had fair function, and 6% had poor function. In 33% of patients (3 cases) with good preoperative hearing level, it was preserved. Newly developed lower cranial nerve dysfunction occurred in 3 patients but proved to be temporary in 2 of them. A CSF leak developed in 6% of those who not previously undergone surgery. Compared with Group B, a significant difference was found only in the rates of the following parameters: excellent facial nerve function, useful and good hearing, lower cranial nerve dysfunction, and blood collection (p < 0.05). The perioperative mortality rate in both groups was 0%.

Conclusions. In patients with a giant VS, total tumor removal can be achieved via the retrosigmoid approach with a 0% mortality rate and low morbidity rate, especially with regards to facial nerve function. In selected cases even hearing preservation is possible. Tumor size significantly correlates with postoperative outcome. (DOI: 10.3171/2009.7.JNS09089)

Key Words • vestibular schwannoma • acoustic neuroma • facial nerve • hearing • functional outcome

Because of the considerable progress in the treatment of VSs in recent decades, complete tumor removal and functional presentation of cranial nerve function is possible in the majority of the cases. However, in cases of large and especially giant VSs measuring > 4.0 cm, the outcome remains less than optimal.\textsuperscript{2,16,17} Although there are a growing number of articles discussing the management of large VSs, few are dedicated specifically to analyzing the outcome after the surgical management of giant VSs.\textsuperscript{5,25,26,30,40} These tumors cause considerable brainstem compression and usually contact and/or compress both the trigeminal and the lower cranial nerves, thus complicating their removal and increasing the intraoperative hazards and the surgery-related morbidity and mortality rates. The best treatment option in such VSs remains controversial; the proposed strategies include partial or subtotal removal followed by observation or radiosurgical treatment of the residual part, or complete removal in 1, 2, or more stages.\textsuperscript{3,14,25} Whatever the treatment choice, preservation of the facial nerve function is complicated due to its significant stretching and extreme vulnerability and even the theoretical possibility of preservation of functional hearing is questioned by some surgeons.\textsuperscript{3,5,41}

The philosophy of the senior author (M.S.) is that in all cases of VS, irrespective their size, complete surgical removal and preservation of facial nerve function should be the goal and—in case functional hearing is present before surgery—an attempt to preserve it should always be made. We analyzed the outcome in a consecutive series of patients with such tumors managed according to this concept. Particular emphasis has been put on surgery-related complication rate, morbidity, and long-term functional outcome. A comparison with a matched group of patients with smaller VSs surgically treated by the same surgeon was performed.
Surgery of giant vestibular schwannoma

Methods

Between 2001 and 2006, > 400 cases of VS were surgically treated at the International Neuroscience Institute in Hannover. In the current study we focus only on those lesions > 4.0 cm. A retrospective chart review of the patients was performed. Patient’s documentation, pre- and postoperative neuroradiological data, operative reports, and follow-up data were reviewed. The following information relevant to the study was recorded: patient age, sex, initial symptoms or signs, neurological status at presentation and after surgery, tumor size, presence of hydrocephalus, surgical complications, and neurological status at discharge and at follow-up. The completeness of tumor removal was assessed both visually at the end of surgery and on postoperative imaging studies.

Included in the study are only those patients in whom the VS was > 4.0 cm in maximal extrameatal diameter. In measuring the size of the VS, we relied on the very strict recommendations of Kanzaki et al. The size of the tumor was determined based on linear planimetric measurements, and only the largest extrameatal diameter was used. The Hannover Classification System for tumor extension was applied, facial nerve function was assessed according to the HB scale, and hearing level was classified according to the New Hannover Classification. Functional hearing levels were considered to be hearing Classes H1 to H2 with pure tone average of up to 40 dB and speech discrimination score > 70%.28

The data obtained in this group were compared with those acquired in a group of patients with VS < 3.9 cm that have been consecutively surgically treated at the same institution.

Outcome Measures and Statistical Analysis

Outcome measures included the completeness of tumor removal, facial nerve function, hearing, and surgery-related complication rate. The chi-square and Fisher exact tests were used to compare the outcome in both groups. A difference at a probability level < 0.05 was accepted as significant.

Patient Population

Group A. Fifty patients fulfilled the criteria for inclusion in the study (Fig. 1A and B). Their mean age was 42.1 years (range 19–73 years); there were slightly more women (52%) than men. The left side was involved in 29 cases and the right side in 21 cases. In 2 patients with neurofibromatosis Type 2, only the giant tumor was taken into account. Ten patients (20%) were previously treated at different institutions: 6 had undergone surgery (1 patient, 7 times; 2, 2 times; and 3, 1 time); 3 had undergone surgical and radiosurgical treatment (1 patient had 2 surgeries prior to radiosurgery); and 1 patient had undergone linear accelerator treatment. In 4 patients a ventriculoperitoneal shunt had previously been placed at another institution. The mean follow-up duration was 34 months (range 5–62 months).

Group B. The group comprised 167 patients with VSs ≤ 3.9 cm. The mean age of these patients was 47.6 years, and 45% were female. The left side was involved in 81 cases and the right side in 86. Five patients had neurofibromatosis Type 2. The mean follow-up duration was 24 months (range 9–45 months). In 13 patients (7.8%) the following treatments had been previously performed: surgery in 10 and surgical and radiosurgical treatments in 3.

In all patients the retrosigmoid suboccipital approach was used with the patient in the semisitting position. Continuous neuropsychological monitoring was performed in all cases and included brainstem auditory evoked potentials, somatosensory evoked potentials, and facial nerve electromyography. The operative technique applied has been presented in detail earlier.27,28

Results

Clinical Presentation

Group A. The most frequent neurological deficit at presentation was hearing dysfunction (Table 1), with some degree of hypacusis being observed in all 50 patients: 4 patients (8%) had normal hearing level (Class 1 of the New Hannover Classification), 6 patients (12%) had useful hearing (Class 2), 9 patients (18%) had moderate hearing (Class 3), 13 patients (26%) had poor hearing (Class 4), and 18 patients (36%) had no hearing (Class 5). Seventeen patients (34%) had tinnitus, and 34 patients (68%) had trigeminal hypesthesia and/or paresthesia: V2 sensory loss in 8 (16%), V3 sensory loss in 6 (12%), V1–3 sensory loss in 11 (21%), and V2–3 in 9 (18%). Two patients (4%) presented with trigeminal neuralgia, in 1 of whom it appeared 36 months after radiosurgery.

Abducent nerve palsy was present in 2 patients (4%), and 5 patients (10%) had low cranial nerve dysfunction. Seven patients (14%) had facial nerve dysfunction: 2 had HB Grade II function, 2 had HB Grade III, 2 had Grade IV, and 1 had HB Grade V. In 4 cases the dysfunction was due to previous surgical or radiosurgical treatment. Gait ataxia was observed in 32 patients (64%), long tract signs in 7 patients (14%), and signs of intracranial hypertension in 13 patients (26%); visual deterioration due to papilledema occurred in 5 patients (10%).

The mean size of the VS was 4.4 cm (range 4–6.5 cm). Radiological signs of hydrocephalus were observed in 16 patients (43.2%).

Group B. The mean size of the VS was 2.3 cm; tumor extension grades are presented on Table 2. In 144 (86%) of 167 patients hearing dysfunction was documented at presentation (Table 1); 14 patients (8%) had complete hearing loss. Tinnitus, vestibular disturbances, and trigeminal hypesthesia were noted in 85 patients (51%), 85 (51%), and 34 (20%), respectively. Nine patients had facial nerve palsy: HB Grade II in 6 patients, Grade III in 2, and Grade IV in 1. Six patients had undergone some form of previous treatment. One patient presented with hemifacial spasm. Radiological evidence of hydrocephalus was demonstrated in 4 patients.

Statistical analysis showed that significant differences existed between the patients in the 2 groups regarding...
the incidence of gait ataxia, long tract signs, and intracranial hypertension.

Surgical Treatment

The VSs were totally removed in all Group A patients (Fig. 1C and D) and in 163 (97.6%) Group B patients. The attempt to dissect the facial nerve from the tumor capsule in 4 patients from the latter group led to intense electromyography changes and, according to the patients wishes, the lesions were subtotally resected, leaving a tiny piece of the tumor capsule attached to the nerve. All 4 of these VSs were large, extending to the brainstem, and 3 of them were cystic.

Facial Nerve Outcome

Group A. The anatomical integrity of the facial nerve was preserved in 46 patients (92%). In the other 4 patients the nerve was found to be extremely atrophic; proximally in the vicinity of the brainstem in 3 and over its entire course in 1 patient (following previous radiosurgery). Five patients had complete facial palsy after surgery, including those with loss of the anatomical integrity of the nerve. Hypoglossal-facial anastomosis was performed in 3 of the 5, whereas the fourth was lost to follow-up. Despite the preservation of the nerve in the fifth patient, it reacted poorly to electrostimulation and a hypoglossal-facial anastomosis was performed 2 weeks later. Follow-up examination showed that facial nerve function recovered to HB Grade IV in 2 patients and to HB Grade III in 2 patients.

At last follow-up 75% of all patients had excellent or good facial nerve function (HB Grade I–III) (see Fig. 3), 19% had fair function (HB Grade IV), and 6% had poor function (HB Grade V). None of the patients had complete facial palsy (Table 3).
Neurological Outcome and Complications

All of them were fed via a gastric feeding tube. In 1 month the dysphagia began to resolve but persisted at the last follow-up 5 months later. He was to be longer. In 1 patient hydrocephalus a temporary external ventricular drain was inserted. One patient (2%) experienced a single epileptic seizure the day of surgery. Cerebrospinal fluid leaks developed in 3 patients (6%) who had not previously undergone surgery: 2 were treated with lumbar drainage and 1 needed surgical closure with removal of the methyImethacrylate cranioplasty. In 1 patient with an epidural air collection the opened mastoid air cells had to be covered with fat tissue and fibrin glue. After surgery, the existing hydrocephalus did not change but was asymptomatic in 4 patients (25%), improved or resolved completely in 11 (69%), and slightly increased in 1 (6%) but needed no further therapy. New hydrocephalus developed in 1 patient and was managed by placement of a lumbar drainage.

The perioperative mortality rate in the group was 0%.

**Group A.** In the patients who had not undergone previous therapy and who useful hearing before surgery, the rate of cochlear nerve preservation was 55.5%; hearing was present in 33% (Table 4) of the patients (Fig. 2). The existing lower cranial nerve dysfunction improved in all 5 patients after surgery. New dysfunction developed in 3 patients but recovered in 2 of the cases. In case of caudocranial nerve dysfunction, oral feeding is avoided until an individual's ability to swallow recovers sufficiently. The third patient had undergone surgery 7 times, and a ventriculoperitoneal shunt had been implanted to manage the hydrocephalus. At admission this patient was in poor general condition, suffering from dysphagia and signs of respiratory insufficiency. A giant cystic VS with largest extrameatal diameter of 6.5 cm was found at surgery and was removed completely. The swallowing problems became aggravated after surgery and a tracheostomy had to be performed because the recovery period was expected to be longer. In 1 month the dysphagia began to resolve but persisted at the last follow-up 5 months later. He was fed via a gastric feeding tube. We observed small blood collections on routine postoperative CT scans in 4 patients—3 in the tumor area and 1 in the cerebellar region (Table 5). All of them were asymptomatic and resolved completely with conservative management. In 1 patient (2%) brainstem and cerebellar edema evolved and a temporary external ventricular drain was inserted. One patient (2%) experienced a single epileptic seizure the day of surgery. Cerebrospinal fluid leaks developed in 3 patients (6%) who had not previously undergone surgery: 2 were treated with lumbar drainage and 1 needed surgical closure with removal of the methyImethacrylate cranioplasty. In 1 patient with an epidural air collection the opened mastoid air cells had to be covered with fat tissue and fibrin glue. After surgery, the existing hydrocephalus did not change but was asymptomatic in 4 patients (25%), improved or resolved completely in 11 (69%), and slightly increased in 1 (6%) but needed no further therapy. New hydrocephalus developed in 1 patient and was managed by placement of a lumbar drainage.

The perioperative mortality rate in the group was 0%.

**Group B.** The overall cochlear nerve preservation was 73.1% (122 patients). If only patients with preoperative hearing and no previous therapy are analyzed, the rate of hearing preservation was 60%. Temporary, mild, low cranial nerve deficit (hoarseness) and transient double vision occurred in 1 patient (0.6%) each. Small blood collections in the tumor area that did not require surgery developed in 2 patients (1.2%). Cerebellar and brainstem edema occurred in 1 patient (0.6%) and was successfully treated by insertion of a temporary external ventricular drain (Table 5). A CSF leak occurred in 4 patients (2.4%): 3 were treated with lumbar drainage and 1 had to undergo surgery. Three of the patients with existing hydrocephalus needed no further treatment. In 1 patient hydrocephalus developed after surgery, and in 1 individual with existing hydrocephalus a temporary external ventricular drain was inserted. Two patients required reoperation for epidural air accumulation, and in 1 the methylmethacrylate cranioplasty had to be replaced.

The tumor recurred 2 years after total VS removal in 1 patient (0.6%).

**Discussion**

The incidence of giant tumors among all cases of VS has been estimated to be up to 2% in Western countries, but in the developing parts of the world they may comprise the majority. In our center they represent 12.5%"
of all surgically treated cases of VS, which might be explained by the referral pattern of the institution.

The clinical presentation of patients with large and giant VSs differs from that in patients with smaller tumors. The cochlear nerve is almost always involved: hearing loss is observed in 96–100% and tinnitus in 42–46%. The trigeminal nerve is the second most frequently involved cranial nerve (in 40–78%), whereas lower cranial nerve dysfunction is noted in 10–14%. The compressive effect on the middle cerebellar peduncle and cerebellum leads to gait instability in 44–88%, and long tract signs due to brainstem compression occur in 14–25%. In our series signs of intracranial hypertension were present in 13 patients (26%) and papilledema with resultant visual deterioration was shown in 5 of these individuals. Thus, excision of the tumor, with decompression of the brainstem and alleviation of the intracranial hypertension, is the mainstay of managing giant VSs.

It is well known that the size of the VS is the main predictor of facial nerve anatomical and functional preservation. As reported by Wiet et al., the risk of facial nerve dysfunction in patients with a VS > 3 cm is 6-fold greater than that in patients with small tumors. The rate of anatomical facial nerve preservation in large VSs is 80–93%, with tumor removal via the translabyrinthine approach or by the retrosigmoid approach. As illustrated by the literature and in the present study, the long-term postoperative facial nerve function in cases of giant VS is worse than that in cases involving tumors < 3.9 cm. In cases of giant VSs, the anatomical integrity of the nerve is preserved in 78–94% and good facial function is preserved in 38–62%. In the current series, excellent or good facial nerve function was documented in 75% of the patients with giant tumors and 91% of those with smaller VSs. However, only the rate of patients with normal function was found to be significantly different in the groups (25% compared with 63%, respectively; p < 0.05).

An important finding of our study is that hearing preservation is possible even in patients with a giant VS. Some authors question even the possibility of preserving hearing in patients with large VSs, and they favor the translabyrinthine approach. However, as correctly pointed by Yates et al., surgeon and patient alike would always choose a hearing-preservation technique if the morbidity were not increased when making the attempt. The low morbidity and complication rates associated with the retrosigmoid approach, the high rate of anatomical preservation of the cochlear nerve, and the chance of hearing preservation support the utilization of this approach even in giant tumors. The comparison of both groups proved that although tumor size is an important predictive factor, if useful hearing is present before surgery, there is still an 11% probability of its preservation.

The structural changes that developed in response to long-standing compression of adjacent neural and vascular structures increase the risk of complications after surgical removal of large and, especially, giant VSs. In a series of 54 patients with VSs > 3 cm, presented by Ro-
land et al.,26 total tumor removal was achieved in 73%. Nevertheless, after surgery 7% of the patients had abduc- cent nerve palsies, 4% had lower cranial nerve dysfunc-
tion, 5% had CSF leaks, and in 6% pseudomeningoele
developed. The incidence of postoperative symptomatic
intracranial hemorrhage in general series is between 1
and 2.4%, but in series of large and giant tumors it in-
creases to 3–4%.16,18,24,31 The major cause of significant
morbidity in the series of Roland et al.26 was postopera-
tive intracranial hemorrhage that had to be evacuated sur-
gically in 4%. Yamakami et al.40 reported that cerebel-
lar hemorrhage resulted in persistent ataxia in 2 patients
(4%). Brennan et al.4 and Glasscock et al.10 indicated that
tumor size is a major predictive factor for the develop-
ment of CSF complications. In series including large or
giant VSs, CSF leakage was the most common surgical
complication, occurring in 4–10%.2,40 Similarly, in our se-
ries CSF leaks developed in 6% of the patients with giant
VS and only in 2.4% of those with smaller tumors, even
though the difference was not significant (p > 0.05).

Different treatment strategies have been put forward
to diminish the operative risks in giant VS and to improve
the functional outcome. Some authors favor subtotal or
radical intracapsular tumor removal to minimize the risk
of injury to the brainstem or the facial nerve.19,26 Staged
removal via a single approach1 or both the retrosigmoid
and the translabyrinthine approaches has been proposed.
in a series of 34 patients treated with the latter strategy,

**TABLE 3: Distribution of HB grades***

<table>
<thead>
<tr>
<th>Facial Nerve Function/HB Grade</th>
<th>% of Patients (no.)</th>
<th>Group A (36)</th>
<th>Group B (145)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>25% (9)†</td>
<td>63% (91)†</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>19% (7)</td>
<td>16% (24)</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>31% (11)</td>
<td>12% (18)</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>19% (7)</td>
<td>8% (11)</td>
<td></td>
</tr>
<tr>
<td>V</td>
<td>6% (2)</td>
<td>1% (1)</td>
<td></td>
</tr>
<tr>
<td>VI</td>
<td>0% (0)</td>
<td>0% (0)</td>
<td></td>
</tr>
</tbody>
</table>

* Patients with preoperative facial nerve palsy and previous treatment were excluded.
† p < 0.05 according to the Fisher exact test.

Patni and Kartush25 achieved total or near-total resec-
tion in all cases without mortality and major morbidity.
In tumors severely compressing the brainstem, radion-
surgery is not an alternative to surgery. However, partial
tumor removal, followed by radiosurgical treatment of the
tumor remnant, is gaining in popularity, although
systematic study of the functional outcome of this treat-
mant in a sufficiently large group of patients has not yet
been performed.14,24 Some modifications of the popular
surgical approaches, such as the combined translabyrin-
thine-retrosigmoid,2 the translabyrinthine-transstentorial
approach,32 the transpetrosal retrosigmoid approach, or
the transmastoid transpetrosal, partial labyrinthectomy
approach,11 claim to offer better access and flexibility in
large and giant VSs. The combined translabyrinthine-
retrosigmoid approach presented by Anderson et al.2 is
presumed to incorporate the strengths of both separate
approaches and, by providing access anteriorly and poste-
riorly to the sigmoid sinus, to minimize the risk of facial
nerve injury. The transapical extension of the enlarged
translabyrinthine approach, introduced by Sanna et al.,30
allows enhanced surgical control over the tumor as well
as the anterior aspect of the cerebellopontine angle in-
cluding the preoptic cistern, and the trigeminal and
abducnt nerves. The translabyrinthine-transstentorial
approach, first described by Morrison and King33 in 1973,
combines the advantages of a translabyrinthine route and
visualization of the tumor from above with the advantage
of early identification of the proximal end of the facial
nerve.32 The translabyrinthine approach offers an excel-
lent anatomical view of the cerebellopontine angle and a direct approach to the tumor that increases the chance of functional preservation of the facial nerve. However, besides being a hearing-destructive approach, in large or giant VS it is associated with a high incidence (14–18%) of CSF leaks.\textsuperscript{6,8,20,32} In the series of Mamicoglu et al.,\textsuperscript{20} CSF leaks occurred in 12 of the patients (17%), 4 of whom required surgery, and meningitis developed in 3 patients (4%). In a series of 190 VSs > 3 cm, Lanman et al.\textsuperscript{18} reported 14% rate of CSF leaks and 4% rate of meningitis.

The advantages of the retrosigmoid approach have been discussed by different authors.\textsuperscript{2,7,8,11,23,40} We also favor this approach and apply it in all patients with a VS. After reviewing our experience, we consider several factors to be of particular importance to reduce surgical morbidity. Both the semisitting position and the constant irrigation by the assistant provide a clear view of the surgical field and enable the surgeon to perform bimanual preparation of the tumor from the surrounding structures. In giant VSs, it might be extremely difficult to identify the arachnoid plane, especially one at the pontomedullary junction. The underlying brain tissue might be noticeably softened, and disrupting the pial plane might lead to brainstem injury and to severe neurological deterioration. The pontine surface veins are a useful guide, and if the tumor capsule alone is held with a forceps and tumor dissection is performed on the level of the arachnoid plane, such inadvertent injuries might be avoided. In large and giant VSs, the draining veins in the region of the cerebellopontine angle might be engorged and extremely fragile, carrying the risk of serious postoperative hemorrhagic complications. We apply jugular venous compression to make any open or torn veins in the cerebellopontine angle visible for final hemostasis twice—at the end of the procedure while the retractor is still in place and after the removal of the retractor to detect bleeding from the supracerebellar bridging veins, if present.

In the present series of giant VSs there were no deaths and there were no significant neurological complications requiring surgical treatment. The existing lower cranial nerve dysfunction improved in all patients after surgery. Although the rate of newly developed lower cranial nerve dysfunction proved to be higher in patients with giant VSs (p < 0.05) it was transient in all cases, improving in 1 and recovering completely in 2 of them. Small asymptomatic blood collections were also seen more frequently in patients with a giant VS (8% compared with 1.2%, p < 0.05), but none required surgical treatment. Ventriculoperitoneal shunt placement before tumor removal has been proposed if symptomatic hydrocephalus is present.\textsuperscript{2} Still, most of the patients recover spontaneously after tumor removal. Of the 16 patients with preoperative hydrocephalus in our series, a slight enlargement was seen in a single patient, but no further therapy was needed. Newly developed hydrocephalus was noted in 1 patient and was managed with temporary lumbar drainage.

**Conclusions**

In giant VSs, total tumor removal can be achieved via the retrosigmoid approach without mortality and low morbidity, especially with regard to facial nerve function. In selected cases even hearing preservation is possible. Tumor size significantly correlates with postoperative outcome.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**


**TABLE 5: New neurological deficits and surgical complication rates**

<table>
<thead>
<tr>
<th>New Neurological Deficit or Op Complication</th>
<th>% of Patients (no.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>lower cranial nerve dysfunction*</td>
<td>Group A (50)</td>
</tr>
<tr>
<td></td>
<td>6% (3)†</td>
</tr>
<tr>
<td></td>
<td>Group B (167)</td>
</tr>
<tr>
<td></td>
<td>0.6% (1)†</td>
</tr>
<tr>
<td>blood collections</td>
<td>8% (4)†</td>
</tr>
<tr>
<td>cerebellar edema</td>
<td>2% (1)</td>
</tr>
<tr>
<td>CSF leak</td>
<td>6% (3)</td>
</tr>
<tr>
<td>epileptic seizure*</td>
<td>2% (1)</td>
</tr>
<tr>
<td>double vision*</td>
<td>0% (0)</td>
</tr>
</tbody>
</table>

* Temporary dysfunction.
† p < 0.05 according to the Fisher exact test.
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